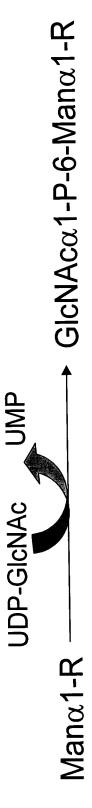


After phosphorylation/uncovering, lysosomal enzyme is recognized by mannose-6-phosphate receptor (M6PR) and targeted to lysosome.



GlcNAc-phosphotransferase

# wild type $\alpha/\beta$ subunit

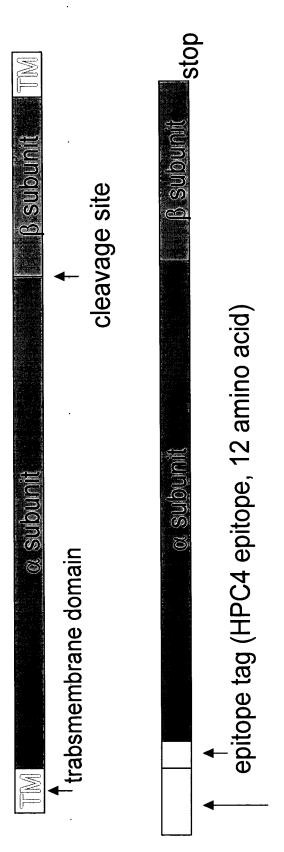


Figure 4

wild type

**X** 

alpha subunit

beta subunit

---DEEESLKTQLAYFTDSKNTGRQLKDTFADSLRYVNKILNSKFGFTS----

---- FTDSKNTG EGRDTFADS----

Factor Xa

---- FTDSKNTDDDDKDTFADS----

Enterokinase

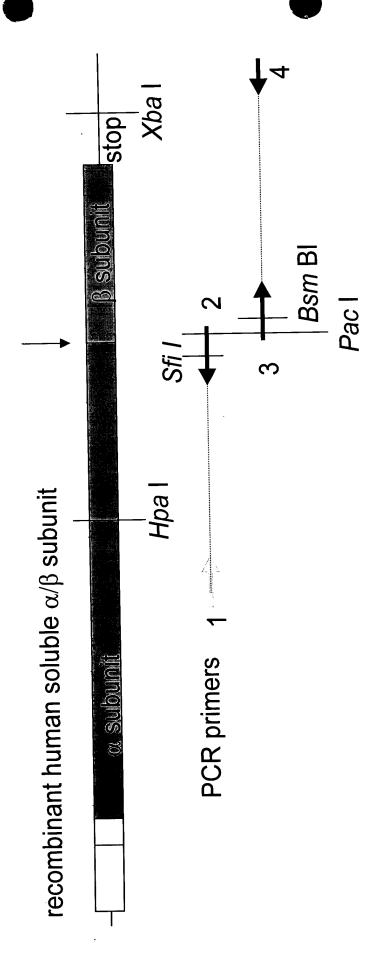
---- FTDSKNRARYKRDTFADS----

Furin

---- FTDSKNPGAAHYDTFADS----

Genease I

Figure 6A



# Figure 6B

5'-GAGGCAATTAATTAACGTCTCTGCAGATTCCCTCAGATATGTAAATAA-3'
S'-GAGGCAATTAATTAACGTCTCTGCAGATTCCCTCAGATATAA-3' 3'-CTCAGTAACTTCTGTGTTAACCGGATGAACCGG<u>AATTAATT</u>CTTATGAC-5'

Pac | BsmB |

PCR primer 2

PCR primer 3

DEEESLKTQLA

A D S L R Y V N K I

Sfil Pac BsmB I

## Figure 6C

---GACGAAGAAGAGTCATTGAAGACACAATTGGCCTACTT GGCCTTAATTAACGTCTCT GCAGATTCCCTCAGATATGTAAATAAAATTC---GCAGATTCCCTCAGATATGTAAAATAAAATTC---TAAGGGAGTCTATACATITATITTAAG--ligation with DNA oligos Sfiland BsmBl BsmB ADSLRYVNKI ---GACGAAGAAGAGTCATTGAAGACACAATTGGCCTACT ---CTGCTTCTTCTCAGTAACTTCTGTGTTAACCGGA DEEESLKTQLAY

---ACACAATTGGCCTACTTCACTGATAGCAAGAATACTGGGATCGAGGGAAGAGATACATTTGCAGATTCCCTCAGA---T Q L A Y <u>F T D S K N T G I E G R D T F</u> A D S L R

Factor Xa

T Q L A Y F T D S K N R A R Y K R D T F A D S L R

T Q L A Y <u>F T D S K N T D D D D K D T F</u> A D S L R

T Q L A Y <u>F T D S K N P G A A H Y D T F</u> A D S L R

TQLAYFTDSKNTGRQLKDTFADSLR

Furin Enterokinase Genease I Wild typee

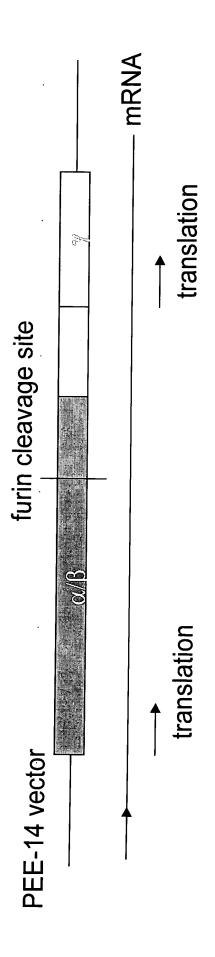


Figure 7